ORIGINAL SCIENTIFIC ARTICLES

Stroke Syndrome: A Case of a Nine Syndrome with a DWI-negative Imaging

Rodelia C. Pascua, MD, MBA[1], and John Harold Hiyadan, MD, FPNA[2]

ABSTRACT

The brainstem contains important structures that give an array of clinical manifestations in pathologic processes. Here, we report a case of "Nine Syndrome" who was admitted in our institution with no acute findings on DWI sequence. The exact prevalence of Nine Syndrome has been accounted to only four cases reported in the literature and this is due to a pontine tegmentum lesion.

Our patient is a 65 year-old male who presented with five-hour history of sudden onset of symptoms presented as ipsilateral gaze palsy, internuclear ophthalmoplegia, a lower motor neuron type of facial palsy, and a contralateral hemiparesis. Using a 1.5T MR cranial scanner and Philips scanner of the time-of-flight of the intracranial vessels, no evidence of acute territorial infarct but an old lacunar infarct was seen in the right pontine area. Both the anterior and posterior circulations are within normal course and caliber with no narrowing seen. Patient was started on dual anti-platelet, high dose statin, and anti-hypertensives on the fourth hospital day.

Nine syndrome is a rare case, and its diagnosis rely on its clinical manifestations, neuroanatomy, and diagnostic imaging. An acute posterior ischemic infarct such as this may yield a negative DWI finding but should not impede the clinician in its early recognition and management.

Keywords: *Nine syndrome, Posterior circulation infarct*

INTRODUCTION

Nine Syndrome is a combination of Eight and a half syndrome plus the unilateral hemiparesis and hemianesthesia or hemiataxia in one case report. The Eight and a half syndrome was first described by Eggenbergerin in 1998 and this was manifested as a combination of one-a-half syndrome characterized by internuclear ophthalmoplegia (INO) caused by a lesion in the medial longitudinal fasciculus (MLF) or paramedian pontine reticular formation (PPRF) and an ipsilateral seventh cranial nerve palsy (1½+ 7 = 8½). These findings combined with a unilateral hemiparesis and hemianesthesia or hemiataxia comprises the Nine Syndrome.

Lesions of the brainstem are now better understood and seen through magnetic resonance imaging. There are only 4 cases reported in the literature pointing to a Nine Syndrome and were mostly attributed to a lesion in the tegmental and ventromedial pons.

OBJECTIVE

To present a rare case of a stroke syndrome: nine syndrome (eight and a half syndrome plus hemiparesis and hemianesthesia) in an adult male, the importance of its early clinical recognition in

[1]Fourth Year resident physician, [2]Consultant, Department of Neurosciences, Baguio General Hospital and Medical Center

correlation of radiologic imaging, and management

CASE PRESENTATION

This is a case of a sixty-five year old male, right-handed who had a five-hour history of sudden onset of dizziness, diplopia when patient looks to his left, with noted frozen right eye, absent horizontal gaze, right-sided facial asymmetry and left-sided weakness and numbness described as dragging of the left foot and poor grip of the left hand. Patient is a hypertensive, non-smoker, non-alcoholic beverage drinker with unremarkable family history. Patient's blood pressure at the ER was 180/110 mmHg.

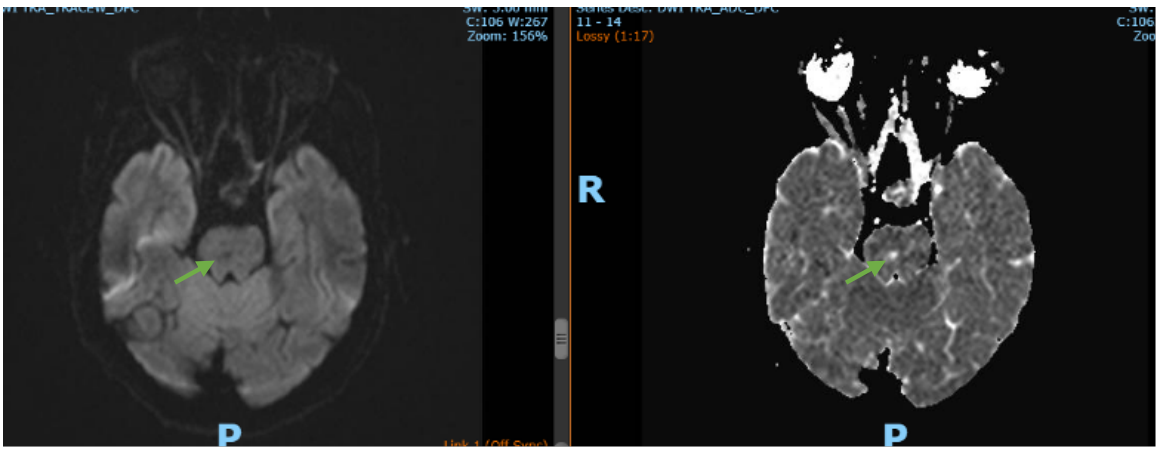
Neurologic examination revealed intact higher cortical functions with a normal mental status examination findings. Cranial nerve examination revealed both pupils that are equally reactive to light, with good direct and consensual reflexes. No ptosis was noted, the left eye was exotropic, confirmed by an asymmetrical Hirschberg test, with no adduction and corrective saccades upon abduction. The right eye was midline on primary gaze, frozen on horizontal gaze; with right-sided peripheral facial palsy and left-sided weakness, with an MMT score of 4/5 and sensory loss of 10%. Cerebellar tests are normal.

Using a 1.5T MR cranial scanner and Philips scanner of the time-of-flight (MRA) of the intracranial vessels, there was no evidence of acute territorial infarct but an old lacunar infarct in the right pontine area (Figure 1). Both the anterior and posterior circulations are within normal course and caliber with no focal dilatation or narrowing seen. Patient was started on dual anti-platelet, high dose statin, and started on anti-hypertensives on the fourth hospital day. He was then sent home improved.

DISCUSSION

Posterior circulation strokes account for 20% of all strokes.[1] In this report, we describe a case of a Nine Syndrome as described in the literature but was noted to have negative acute findings on MRI-DWI sequence. To date, there are only four reported cases of Nine Syndrome. Three reported cases of Nine Syndrome involved pontine infarcts in the tegmental, ventromedial, ventrolateral medipeduncle area and a pontine hemorrhage extending from the pontine tegmentum to the anteromedial, dorsolateral, and above the pons.[2,3,4,5] The fourth case report showed two acute infarcts in the right paramedian pons and dorsal pons.[6] All cases reported clinical manifestations of Eight-and-a-half syndrome

Figure 1. MRI DWI with the corresponding ADC sequence showed a chronic infarct (green arrow) at the right pontine tegmentum with



with additional findings invoking the corticospinal tract and medical lemniscus. One case reported by Mahale et al showed contralateral hemiataxia.[4]

With the advent of MRI, we now better understand the localization of brainstem lesions and its correlation with our clinical findings. However, in this report, MRI findings revealed no evidence of acute territorial infarct or bleed, but an old lacunar infarct in the right pontine tegmentum. Edlow et al in their meta-analysis showed a pooled prevalence of DWI-negative acute ischemic stroke of 6.8%, 95% confidence internal (CI) 4.9-9.3 and is strongly associated with posterior circulation ischemia (OR 5.1, 95% CI, 2.3-11.6, p <0.001). [7]

CONCLUSION

Nine syndrome is a rare case, and its diagnosis relies on its clinical manifestations, neuroanatomy, and diagnostic imaging. An acute posterior ischemic infarct such as this may yield a negative DWI finding but should not impede us, the clinicians, in its early recognition and management.

REFERENCES

1. Merwick A, Werring D. Posterior circulation ischaemic stroke. BMJ. 2014 May 19;348:g3175. doi: 10.1136/bmj.g3175
2. Fang, et al. One-and-a-half syndrome with its spectrum disorders. Quantitative Imaging in Medicine and Surgery 2017; 7 (6): 691-697
3. Rosini F, Pretegiani E, Guideri F, Cerase A, Rufa A. Eight and a half syndrome with hemiparesis and hemihypesthesia: The nine syndrome? Journal of Stroke Cerebrovascular Disease. 2013; 22:e637-8
4. Mahale RR, Mehta A, John AA, Javali M, Abbas MM, Rangasetty S, et al. "Nine Syndrome: A new neuro-ophthalmologic syndrome: Report of two cases. Ann Indian Acad Neurol. 2015;18:335-7
5. Yong Muh NG, Kang PS, Khor CG, Ping Seung ON, Letchuman G. "Nine" syndrome and its MRI brain findings. Neuroly Asia 2017;22:349-52
6. Singhdev, Jennifer, et al. Nine Syndrome: Case Report and Review of Clinical Signs in Internuclear Ophthalmoplegia
7. Edlow B, Hurwitz S, Edlow J. Diagnosis of DWI-negative acute ischemic stroke: A meta-analysis. American Academy of Neurology. 2017; 89(3): 256-262